Lesson 235: PreAnesthetic Assessment of the Patient With Dermatomyositis

PREANESTHETIC ASSESSMENT

A COURSE OF STUDY FOR AMA/PRA CATEGORY 1 CREDIT

1) Read this article, reflect on the information presented, then complete the lesson quiz and course evaluation. Return it to Mount Sinai School of Medicine, Department of Anesthesiology, before November 30, 2005. (CME credit is not valid past this date.)

2) You must achieve a score of 80% or better to earn CME credit.

3) The estimated time to complete this activity is 2 hours.

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NEEDS STATEMENT

Dermatomyositis (DM), which has the highest incidence rate of all the idiopathic inflammatory myopathies, is potentially treatable. Although DM is not very common, it is necessary for the clinical anesthesiologist to recognize the signs and symptoms of the disease because of the variable manifestations and broad systemic implications of untreated DM. If the patient’s muscles of respiration are severely affected, the anesthesiologist must be prepared to consider periphereic assisted ventilation. In addition, the anesthesiologist must consider the possible presence of myocardial dysfunction, anemia, interstitial pneumonia, or occult carcinoma when selecting anesthetics and neuromuscular blockers for the patient with DM.

CASE HISTORY

A 22-year-old man, scheduled for elective hernia repair, presented with Raynaud’s phenomenon, a generalized rash on his face, and violaceous plaques around his elbows and interphalangeal joints. On examination, the plaques were found to be Gottron’s papules. Periarticular edema and a heliotrope rash of the upper eyelids were also noted. The patient stated that these symptoms had persisted for more than 6 months, and that he was finding it increasingly difficult to perform tasks requiring exertion. A review of systems revealed mild to moderate dyspnea. Tests of muscle strength indicated significant deficit of proximal muscle function in the upper and lower extremities. Laboratory test results showed elevated serum levels of aldolase (10 U/L) and creatine kinase (250 U/L). The results of further tests were negative for human immunodeficiency virus and hepatitis B virus. A chest radiograph revealed mild interstitial disease. Muscle biopsy specimens of the left triceps showed perifascicular atrophy, peryvascular inflammation, and sparing, but significantly dilated, capillaries within the muscle fiber. Additional results of routine laboratory and physical examinations were within normal limits.

TARGET AUDIENCE

Anesthesiologists

SIGN AND SYMPTOMS

Dermatomyositis shares many symptoms with polymyositis and inclusion body myositis. Common clinical presentations include atrophy of the muscles of the extremities and accompanying weakness. If left untreated, the disease can progress to advanced stages in which systemic involvement ranges from restrictive pulmonary defects to cardiomyopathy. Hallmark skin lesions differentiate DM from other idiopathic inflammatory myopathies: patients with polymyositis or inclusion body myositis do not exhibit cutaneous manifestations. The characteristic heliotrope rash of DM often occurs on the face, neck, and back. Also present is the Gottron rash—a scaly, violaceous papule. Further differentiating DM from other idiopathic inflammatory myopathies is an autoimmune process that is particularly devastating to muscle fibers. A muscle biopsy specimen of an affected patient often reveals perifascicular atrophy, hypoperfusion, and fibrosis of muscle fibers resulting from continual degeneration and regeneration. This troubling muscle necrosis is due to a decreased density of the endofascicular vasculature, leading to ischemia in distant fibers. Complement factors activate B cells and CD4+ T cells against the capillaries, which leads to damage of the microvasculature. However, a recent report suggests that perifascicular atrophy is a rare finding in adults with DM and appears very late in the disease. Instead, early diagnostic features of a DM muscle biopsy specimen are thought to be deposits of the membranolytic attack complex and tuboreticular inclusions within the endothelium.

Classification

In 1975, a paper by Bohan and Peter suggested 5 classes of idiopathic inflammatory myopathy and listed diagnostic criteria for each. Although it has been made obsolete by newer technologies and recent discoveries about the disease, the older scheme is still used by many as the basis for categorizing patients. The 5 classes of idiopathic inflammatory myopathy proposed by Bohan and Peter are the following:

1) Primary, idiopathic inflammatory polymyositis
2) Primary, idiopathic DM
3) DM—polyomyositis associated with neoplasia
4) Juvenile DM—polyomyositis associated with vasculitis
5) DM—polyomyositis with associated collagen-vascular disease

Some scientists consider this classification scheme outdated because it does not account for many cases of DM. For example, patients who have amyopathic DM present with DM-like lesions, but without the extensive systemic and muscle involvement of classic DM. Also, until the publication of a paper by Sayers and colleagues in 1992, inclusion body myositis was not recognized see Lesson 235 page 42
Table 1. Dermatomyositis: Subsets and Definitions

<table>
<thead>
<tr>
<th>Terms (abbreviation)</th>
<th>Definitions</th>
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<tr>
<td>Amyopathic DM (ADM)</td>
<td>A subtype of DM in which patients have biopsy-confirmed hallmark cutaneous manifestations of classic DM for 6 months or longer but no clinical evidence of proximal muscle weakness and no serum muscle enzyme abnormalities. If more extensive muscle testing is carried out, the results should be within normal limits. (If such results are positive or abnormal, the disease can be classified as hypomyopathic DM.) Exclusion criteria for amyopathic DM include the following: 1) Treatment with systemic immunosuppressive therapy for at least 2 consecutive months within the first 6 months after onset of skin disease; such therapy can prevent the development of clinically significant myositis. 2) Use of drugs known to be capable of producing isolated DM-like skin changes (eg, hydroxyurea, statin cholesterol-lowering agents) at the onset of cutaneous DM changes.</td>
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<td>Classic DM (DM)</td>
<td>Patients have the hallmark cutaneous manifestations of DM, proximal muscle weakness, and objective evidence of the characteristic muscle inflammation of DM.</td>
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<tr>
<td>Hallmark cutaneous lesions of DM</td>
<td>Skin lesions that alone or in combination are seen only in patients with some form of DM (synonymous with DM-specific skin disease).</td>
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<tr>
<td>Hypomyopathic DM</td>
<td>Patients with DM-specific skin disease and no clinical evidence of muscle disease show subclinical evidence of myositis on laboratory, electrophysiologic, or radiologic evaluation.</td>
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<tr>
<td>Idiopathic inflammatory dermatomyositis (IIDs)</td>
<td>A more inclusive designation for the spectrum of illnesses that has conventionally been referred to as idiopathic inflammatory myopathies.</td>
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<tr>
<td>Juvenile DM (JDM)</td>
<td>DM with onset in childhood, commonly characterized as a vasculitis. Patients with JDM are at greater risk for the development of calcinosis.</td>
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DM, dermatomyositis
Adapted from: Sontheimer.2

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as a separate category of idiopathic inflammatory myopathy.9 Other classifications currently proposed include drug-induced DM and hypomyopathic DM. The different forms of DM currently accepted by most physicians are listed in Table 1.

Epidemiology

A study by Chen et al in 2001 showed that, of all the inflammatory myopathies, DM has the highest prevalence rate among adults and children.8 Data on incidence rates are relatively scarce because of the perception that DM is a rare disease. Varying numbers have been reported, typically fluctuating between 2.2 and 10 cases per million. The fluctuation depends on the region in which the study was performed, the time period of the study, and the criteria used to diagnose and differentiate between myopathies.10,11 Until recently, cases of amyopathic DM were not included in the statistics for DM.2

The onset of DM is gradual, although the skin lesions can be a harbinger of developing muscle weakness; hallmark cutaneous lesions typically present weeks to months before the onset of muscle disease. The proximal muscles of the limbs are preferentially affected, causing difficulty with everyday tasks and movements. Most patients become symptomatic in their 50s or 60s.12 However, the prevalence of childhood-onset DM has led scientists to create a classification known as juvenile DM.

The regional variability of the results of epidemiologic studies has made it difficult to generalize disease rates (eg, gender-specific). Studies from Singapore, Taiwan, and Hong Kong have reported higher incidence rates in males than in females. However, studies conducted in North American and Israeli populations have reported twice as many occurrences of the disease in females as in males.13,14 The matter is further complicated by the results of specific breakdown analyses of populations. For example, in a study conducted by Oddis and colleagues, African-American women were found to be more susceptible to DM during their childbearing years.15

The incidence rate for juvenile DM has been estimated at 3.1 per million children per year, according to a National Institutes of Health registry.16 As in adult DM, the female-to-male ratio in juvenile DM is 2:1 in North American population studies, whereas an Asian study reported a higher prevalence in boys.17 Symptoms usually develop in children between their 6th and 12th years, with a mean age at onset of 7 years. Juvenile patients with DM are more likely to have a vasculopathy that eventually develops into dystrophic calcification—an occurrence rarely seen in the adult form of DM.17 Environmental factors have been found to be associated with the prevalence of DM; seasonality has been suggested as a related factor in both adult and juvenile DM.3-9 Geographic clustering has also been demonstrated in studies conducted in Europe and North America, possibly as a result of the transmission of a genetic predisposition.20,21 Exposure to ultraviolet light, primarily UV-A and UV-B rays, has triggered DM in some patients.22 Other factors that may activate DM include vaccine and drug exposure.23

It has been difficult to document the prevalence of clinical amyopathic DM because of the use in many studies of the criteria of Bohan and Peter, which do not account for amyopathic DM.7 Although it was originally thought to be a rare disease, more than 300 case studies of amyopathic DM have been published since its recognition. A European paper reported that 10% of all cases originally diagnosed as DM were eventually found to better fit the criteria for amyopathic DM.24 A Taiwanese study found an incidence of amyopathic DM of approximately 14% among all diagnosed cases of DM; consistent with the trend in Asians, the incidence of amyopathic DM was higher in males than in females. The study reported that the frequency of amyopathic DM was second only to that of adult-onset DM, so it is more common than originally believed.9

Pathology

Although no specific antigens have been shown to cause DM, the general hypothesis is that environmental agents evoke a humoral autoimmune response against the body’s own tissues, which leads to self-intolerance. The complement-mediated process includes capillary necrosis, resulting in hypoperfusion of muscle fibers. Cutaneous lesions stem from complement deposition along the dermal–epidermal junction.2

Several studies have suggested a genetic predisposition in patients with DM. One study has linked DM to the 8.1 ancestry haplotype; specific alleles that predispose to autoimmunity production, and consequently DM, include HLA-B8, HLA-DR3, and HLA-DQ2.25 Also, a more recent study has linked a mutation in the tumor necrosis factor-α (TNF-α) promoter TNF-308A to disease symptoms.26 Studies of patients with juvenile DM suggest that the HLA-DQA1*0501 allele plays an important role but may not be solely responsible for all facets of the disease.27 Other genes thought to play a role include the HLA class II alleles DRB1*0301 and DQA1*0501. Another study found that genetic factors affecting Japanese patients were different from those affecting Caucasian patients; patients in Japan were more likely to carry the DRB1*0701 allele and were less likely to be affected by the DRB1*0301 and DQA1*0501 alleles.28

A genetic predisposition is typically not enough to cause a patient to be symptomatic; an environmental trigger often plays an important role in the loss of self-tolerance.29 Exposure to ultraviolet light has been studied as a causative factor in patients with dermal lesions. Exposure to UV-A and UVB radiation leads to elevated levels of TNF-α—seemingly resulting from increased production by epidermal keratinocytes.23 This polymorphism causes increased apoptosis of keratinocytes, which in turn produces the photodistribution of lesions seen in DM patients after extended exposure to UV radiation.27 Exposure to infections also can lead to an autoimmune response. Although the involvement of coxsackievirus and human immunodeficiency virus (HIV) has been suggested, studies have been unsuccessful in isolating viral genome from affected muscle and have not proved that contact with these antigens leads to a loss of self-tolerance.29

The autoimmune response includes the production of autoantibodies. Each specific clinical syndrome is associated...
with an autoantibody, although the mechanisms are often similar. It is difficult to use autoantibodies as a diagnostic tool in DM because the prevalence of any single antibody is only 20% in DM patients. Two general classes of autoantibodies are involved in DM: antibodies targeted against nuclear antigen inhibit protein synthesis, resulting in anti-synthetase syndrome, whereas antibodies targeted against cytoplasmic antigen prevent translational transport. Autoantibodies of the latter class recognize the signal recognition particle (SRP); patients with this autoantibody are at greater risk for cardiac complications and do not respond well to known therapies.

The autoantibodies that inhibit protein synthesis—for example, Jo-1, PL-7, PL-12, and OJ—are highly specific for DM; each recognizes a specific transfer ribonucleic acid (tRNA) synthetase as its antigen. For example, Jo-1 carries molecular specificity for histidyl-tRNA synthetase, and PL-7 carries specificity for theophylline-tRNA synthetase. The result is “anti-synthetase syndrome,” which is prevalent in many cases of DM. Antisynthetase syndrome is associated with Raynaud’s phenomenon (arterial contraction, typically in the digits) and an increased risk for arthritis and interstitial lung disease. In contrast, amyopathic DM does not affect the musculature. Thus, the autoantibodies produced by patients with amyopathic DM differ from the tRNA synthetase and SRP autoantibodies produced by patients with myopathic DM. A recent study suggested that patients with amyopathic DM preferentially produce autoantibodies of the 155 kDa and Se antigens.

Injury to the musculature commences when autoantibodies, directed against the capillary endothelium, activate complement factor C3. A cascade of events through the lytic component of the complement pathway—that is, the production of complement factors C3b and C4b—results in the deposition of membrane attack complex in the endothelial lining of muscle capillaries. The endothelial cells subsequently swell and necrose, causing inflammation. As the capillary walls are destroyed, the muscle fibers are deprived of blood flow and become ischemic. In the patient with DM, the sharp decline in the ratio of capillaries to muscle fibers causes injury on a more chronic level. If the blood flow from increasing, as would regularly occur during activity, the muscle fibers swell and necrose, causing inflammation. As the disease progresses, the muscle fibers are more susceptible to damage in DM, and perifascicular spaces. B cells and CD4+ T cells are found most commonly, supporting the view that the autoimmune process is humorally mediated. Damaged fibers express the major histocompatibility complex class I antigen, intercellular adhesion molecule, and neural cell adhesion molecule, particularly in the perifascicular regions.

Cutaneous lesions show epidermal atrophy from perivascular inflammation; CD4+ T lymphocytes are commonly found in the dermis. Chronic inflammation leads to degeneration of the basement membrane and vacuolization of the basal keratinocytes. Dermal changes include interstitial mucin deposition and some lymphocytic infiltrate. A biopsy specimen around the characteristic Gottron’s papules typically shows acanthosis.

**Systemic Manifestations**

As its name suggests, skin lesions are one of the major signs of DM. In a recent review, Sontheimer suggested major and minor criteria for classifying all skin lesions of DM (Table 2). The major criteria are characteristic of DM, and the presence of any 2 major criteria indicates a cutaneous manifestation of DM. In patients with DM, changes in the skin occur either gradually or with rapid onset. The slower form tends to progress as transient erythematous rashes that are randomly distributed on the body; these gradually evolve into characteristic Gottron’s papules and a heliotrope rash. Patients with acute-onset DM have persistent erythematous rashes; the lesions usually present in a photodistribution that is characteristic of DM.

Change to the musculature is the other major effect in patients with DM. The autoimmune attack on myocytes preferentially affects proximal muscle groups in a symmetric pattern; however, as the disease progresses, all muscles are susceptible. Patients often report fatigue and malaise. More serious cases, patients are unable to complete their activities of daily living because of severe muscle weakness. Although it would seem logical, the level of weakness does not necessarily correlate with the degree of inflammation or damage seen in a muscle biopsy specimen. Sontheimer has suggested that high levels of cytokines may aggravate symptoms of muscle weakness without producing a characteristic myopathic inflammation.

It should be noted that cutaneous DM lesions can occur without muscle involvement, as in patients with amyopathic DM. Yet another form of DM, known as hypomyopathic DM, mimics amyopathic DM clinically but is characterized by laboratory findings in muscle biopsy specimens. Thus, the degree of cutaneous and muscle involvement is unique to each patient.

Dermatomyositis can cause other systemic problems, including an increased risk for malignancy. It is important for clinicians to quickly recognize the signs of DM and diagnose the disease in its early stages so that further investigations may be undertaken. A cohort study found that patients in whom DM was diagnosed were 3 times as likely to have a malignancy within the first 5 years after the initial diagnosis; it has also been found that ovarian cancer is almost 10 times more likely to develop in women with DM than in controls. Advanced age and relatively severe cutaneous and muscle involvement seem to correlate with a greater risk for cancer; however, complications of DM (eg, interstitial lung disease) have not been similarly linked. It has been reported that females with DM are at higher risk for cancer than males with DM; however, gender-specific incidence rates are controversial, and some reports have found higher rates in men than in women. A population-based study found that patients with DM were at greatest risk (from highest to lowest) for ovarian, lung and pancreatic cancer; non-Hodgkin’s lymphoma, and gastric

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**Histopathology**

A muscle biopsy is one of the most important processes in diagnosing DM. A typical biopsy specimen will show inflammation around the fascicles within the perivascular space. As a result, atrophy around 3 or 4 layers of the fascicle, due to myocyte necrosis and microinfarcts, is generally seen. Vacuolization of the fascicles is also evident, as muscle fibers around the damaged capillaries lose their source of nutrition. A reduced density of capillaries (as a result of the attack on endothelial cells) can also be observed. Type II muscle fibers tend to be more susceptible to damage in DM, and regeneration often yields hypertrophic tissue with centralized sarcoclemmal nuclei.

Lymphocytic infiltrate can also be found in the perivascular and perifascicular spaces. B cells and CD4+ T cells are found most commonly, supporting the view that the autoimmune process is humorally mediated. Damaged fibers express the major histocompatibility complex class I antigen, intercellular adhesion molecule, and neural cell adhesion molecule, particularly in the perifascicular regions.

Cutaneous lesions show epidermal atrophy from perivascular inflammation; CD4+ T lymphocytes are commonly found in the dermis. Chronic inflammation leads to degeneration of the basement membrane and vacuolization of the basal keratinocytes. Dermal changes include interstitial mucin deposition and some lymphocytic infiltrate. A biopsy specimen around the characteristic Gottron’s papules typically shows acanthosis.
Table 2. Proposed Minimal Set of Hallmark Cutaneous Manifestations of DM for Defining Amyopathic DM in Clinical Studies

<table>
<thead>
<tr>
<th>Major Cutaneous Criteria</th>
<th>Minor Cutaneous Criteria</th>
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<tr>
<td>• Heliotrope rash: macular violaceous erythema, with or without associated scale of the eyelids or periorbital skin. Secondary or associated cutaneous findings (eg, scale, pigmentary change, telangiectasia), or edema may also be present.</td>
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<td>• Gottron’s papules: violaceous papules or small plaques overlying the dorsal or dorsolateral aspects of interphalangeal or metacarpophalangeal joints. When fully formed, the papules become slightly depressed at the center, which can assume a white, lacy appearance. Associated scale–hyperkeratosis, pigmentary change, or telangiectasia may be present.</td>
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<tr>
<td>• Gottron’s sign: macular violaceous erythema, with or without associated scale–hyperkeratosis, pigmentary change, or telangiectasia involving extensor aspects of the knuckles, elbows, and knees or medial malleoli.</td>
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<tr>
<td>• Periungual nail-fold telangiectasia or cuticular hemorrhage–infarct, with or without dystrophic cuticles</td>
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<tr>
<td>• Poikiloderma (concurrence of hyperpigmentation, hypopigmentation, telangiectasia, and superficial atrophy)</td>
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<tr>
<td>• Prurigo: pruritus, eczematous lesions. Alcohol has been found to cause DM-like cutaneous eruptions.</td>
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and colorectal cancers. Regional studies have indicated a greater risk for nasopharyngeal cancer in Asian patient.12,17

Respiratory system changes frequently occur in patients with DM. The primary cause is the autoimmune attack on the musculature: the muscles of respiration become weak, and a restrictive defect is observed. The diffusion capacity is reduced, and patients experience exertional dyspnea. The production of nuclear autoantibodies—concurrently leading to antiphospholipid syndrome—also increases the risk for pulmonary complications and makes patients more susceptible to opportunistic infections and interstitial lung disease (observed in as many as 80% of DM patients). In cases of interstitial lung disease with an acute onset, patients deteriorate quickly, and many are resistant to treatment; they can die within a month after death.16

Dermatomyositis can affect many other systems in the body. For example, patients who produce the anti-SRP antibody are thought to be at greater risk for cardiomyopathy. However, 2 recent studies noted a lack of cardiac involvement in patients with anti-SRP antibody. Cardiac and systemic problems can also develop as a result of the long-term use of steroid drugs to treat other symptoms of DM. The autoimmune attack on muscles can cause dysphagia due to weakened upper esophageal contractions, which increases the risk for aspiration. Calcifications can occur, particularly subcutaneously and within muscle tissue. If calcifications are untreated or unre sponsive to treatment, ulcerations, tenderness, and pain can develop in affected areas.14

Diagnosis

A correct diagnosis of DM is difficult to make because patients with other autoimmune diseases present with similar symptoms. A skin biopsy of suspected cutaneous lesions can provide evidence of the onset of the disease; the patient must be rigorously screened for muscle and systemic involvement. In general, the following tests can confirm the onset of myopathy: 1) measurement of serum muscle enzyme concentrations; 2) electromyography (EMG); and 3) muscle biopsy.15

Creatine kinase (CK) is the major muscle enzyme evaluated in suspected cases of DM. As the most sensitive enzymatic parameter, the CK levels can be increased to up to 50 times normal levels in a patient with DM. Jorizzo has estimated that 95% of patients have elevated CK levels at some point during disease progression.16 The isoenzyme that usually contributes most to the elevated enzyme levels is CK-MM, but the CK-MM isoenzyme has also been found in the serum of patients who do not exhibit cardiac complications.17 Typically, CK levels correlate with the level of disease activity, although recent studies have also found patients with normal CK levels. Levels of other enzymes are found to be elevated as the result of their release from damaged muscle tissue. The serum myoglobin concentration is increased, even in the early stages of muscle degeneration. Elevated concentrations of aspartate transaminase, alanine transaminase, and lactate dehydrogenase can be detected as the autoimmune response becomes more aggressive.18

Electromyography is another test that can verify suspected cases of DM. More than 90% of patients with DM have abnormal electromyograms. Needle EMG shows fibrillations and increased spontaneous activity. Systematic repetitive discharges and positive sharp waves are also common.19 The presence of EMG findings can confirm, but does not necessarily indicate, a diagnosis of DM. Many other myopathies, such as polymyositis, can generate similar findings. The physician must be wary in making a diagnosis of DM based primarily on needle EMG.20 Perhaps the most important laboratory test for diagnosing DM is the muscle biopsy. Because axial muscles are affected first, it is recommended that the biopsy specimen come from the triceps. Findings should correlate with the pathology of DM—that is, inflammation in the perivascular and perifascicular space.21 Fiber necrosis should be located on the periphery of the fascicle, but dying myocytes may be found in the middle, where capillaries have been destroyed. The attack on endothelial cells results in a dramatic decrease in the capillary-to-fiber ratio, and residual capillaries are dilated to compensate for the decreased blood flow.

Other tests that have been conducted in certain groups of patients do not provide good diagnostic criteria for the general population of patients with DM. The erythrocyte sedimentation rate is elevated in 50% of patients; rheumatoid factor has been found to be elevated in 20% of patients. One study has found increased levels of von Willebrand factor and CD19+ B cells.22 It has also been reported that major histocompatibility complex class I antigen was expressed in two thirds of a sample of patients.

Magnetic resonance imaging and ultrasonography have proved beneficial in diagnosing the disease. Ultrasonography has helped determine sites for muscle biopsy and, with its relatively low cost, can be used in a large number of patients. Although magnetic resonance imaging is more expensive, it is one of the most sensitive tests for myopathy.23

The diagnosis of DM can be confusing because patients exhibit different levels of cutaneous and muscle involvement, and some may present with signs overlapping with those of other connective tissue diseases. Early signs of DM can be mistakenly attributed to contact dermatitis, psoriasis, scleroderma, or atopic dermatitis. In more advanced stages, DM can be easily mistaken for lupus erythematosus. Most prominently, the hallmark skin lesions of DM, Gottron’s papules, can rule out a diagnosis of lupus erythematosus. Cutaneous manifestations are more commonly “violaceous” in DM, whereas the lesions of lupus are reported to appear as more red or pink. Laboratory tests can also help with the differential diagnosis; patients with lupus erythematosus usually test positive for anti–double-stranded DNA and anti-Sm antibodies, whereas such findings are absent in DM.24

Diseases that do not affect collagen tissue can sometimes mimic DM. The physician must be careful to rule out certain viruses that can cause myopathies, such as HIV, Epstein–Barr virus, coxsackievirus, and hepatitis B virus. Neurologic disorders can also present with proximal muscle weakness similar to that seen in DM. Myasthenia gravis, Guillain–Barré syndrome, and Lou Gehrig’s disease (amyotrophic lateral sclerosis) can mislead the physician when other cutaneous findings are present. In addition, many drugs can trigger cutaneous lesions. Alcohol has been found to cause DM-like symptoms in certain patients; corticosteroids can also cause cutaneous eruptions.25

Treatment

The treatment of DM focuses on 2 aspects of the disease—the cutaneous lesions and the systemic effects. The local application of superpotent corticosteroids and glucocorticoids can help alleviate inflammation and pruritus; these medications must be used cyclically in long-term treatment. Most other treatments (eg, shampoo for the scalp, gels for problem spots) are supportive, to relieve itching. Antihista mines can also be used to counter minor episodes of pruri tus throughout the day.26 The patient must be vigilant in applying sunscreen and avoid prolonging, unprotected exposure to the sun. As seen by the cholinergic response, UV radiation can trigger DM by prompting autoimmune changes at the polymorphic in the TNF-308A allele.27 Every care must be taken to eliminate further provocation of the autoimmune response.

Most treatments focus on restoring muscle strength and relieving other manifestations, thus restoring normalcy as much as possible to the patient’s life. Although corticosteroids are typically thought of as the first line of defense, the use of nonsteroidal solutions should be maximized before the...
patient is subjected to the many side effects of steroids. Hydroxychloroquine sulfate and chloroquine have been suggested for treating the cutaneous manifestations of the disease; however, the patient must be closely monitored for signs of renal toxicity. 

Dapsone, a drug typically used to treat leprosy, was found to be of value in a recent case study of 2 patients with disease resistant to other therapies. 

Prednisone is the glucocorticoid used most often for treating DM. Studies have shown a better prognosis, greater recovery of muscle strength, and a reduction in cutaneous signs after 1 to 3 months of treatment. The regimen should be administered for at least 1 year to prevent a recurrence; during this time, the dose can be gradually decreased to minimize side effects. 

Patients who cannot take prednisone for various reasons, find its side effects unacceptable, or do not respond to it may be given immunosuppressants. Azathioprine has shown efficacy after 4 to 6 months. Methotrexate acts faster than azathioprine but may cause pneumonitis. The decision to use an immunosuppressant and the selection of a specific agent depend on the profile of the patient. Benefits and risks should be evaluated before the therapy is administered. 

Other proposed treatments have included plasmapheresis, anti–TNF-α therapy, and I.V. immunoglobulin. A study of I.V. immunoglobulin indicated that the therapy seemed to resolve the immunopathology of DM. However, the effects of treatment were short-lived, and intusions were required every 2 months. 

Most treatments cause enzyme levels to quickly return to normal. The physician must be cautious about pursuing normal laboratory test results. However, the drugs can help lower serum enzyme levels without actually affecting the disease state; some reports even describe patients who presented with normal CK levels throughout the course of the disease. Instead, the alleviation of cutaneous symptoms and improvement of muscle strength are the goals of steroid therapy, and the physician should monitor the patient for changes or improvement in that regard.

Anesthetic Considerations 

The patient with DM must be evaluated preoperatively for muscle weakness. If the muscles of respiration are severely affected, the anesthesiologist should consider assisted ventilation throughout surgery and even postoperatively, possibly for an extended length of time. 

An assessment of head and neck mobility can determine whether awake fiberoptic laryngoscopy and intubation are necessary in the presence of a DM. 

Neck mobility can determine whether awake fiberoptic laryngoscopy and intubation are necessary in the presence of a DM. 

Plasmapheresis decreases the concentration of circulating antibodies. Thus, muscle relaxants such as succinylcholine have been used in the presence of a DM. 

DM may be unduly sensitive to regular doses of anesthetic drugs. Concurrent treatments for DM can alter drug sensitivities. The systemic effects of DM include anemia and interstitial pulmonary edema. An active myositis may attack the striated muscle of the upper esophagus, causing dysphagia and gastroesophageal reflux. Patients are at risk for aspiration pneumonia, and prophylaxis is warranted. Myocardial dysfunction should be considered when anesthetics are selected. 

The occipitocervical muscles frequently detect in patients with DM may be associated with neuromuscular weakness and myasthenic syndrome. It has been suggested that doses of relaxants be titrated carefully. 

Neuromuscular block monitoring is essential. 

Because of their decreased muscle mass, patients with DM may be unduly sensitive to regular doses of anesthetic drugs. Thus, muscle relaxants such as succinylcholine have traditionally been avoided in these patients. 

One case study, however, reported the successful use of succinylcholine in a patient with DM. It must be noted that the patient exhibited a response similar to that of patients with myotonic dystrophy—that is, a short-lived contracture response. 

Concurrent treatments for DM can alter drug sensitivities. 

Plasmapheresis decreases the concentration of circulating serum cholinesterase. Because this enzyme breaks down exogenous esters and inactivates drugs commonly used in anesthesia, a lack of cholinesterase can lead to prolonged relaxation.

Management of the Case 

Dermatomyositis was diagnosed in this patient, and the elective surgery was postponed. Prednisone was prescribed in divided doses of 1.5 mg/kg per day. The patient was referred for screening for possible malignancies. At a follow-up visit 2 weeks later, the CK and aldolase levels were still high but within normal limits. Two months after starting treatment, the patient demonstrated significant signs of improvement; the cutaneous lesions had diminished, and muscle strength was returning. A muscle biopsy showed regenerating muscle fibers and an increase in the ratio of capillaries to muscle fibers. The administration of prednisone was consolidated into 1 daily dose and slowly tapered over the course of 12 months.

Summary 

If left untreated, DM can be debilitating. The effect of the autoimmune response on the musculature causes severe muscle weakness and impedes the patient from performing daily activities. Because of this devastating effect, it is important for the clinician to recognize signs early on and start treatment as soon as possible. The patient should also be educated about potential disease triggers and take precautions, such as avoiding extended exposure to UV irradiation. 

Anesthesiologists should be keenly aware of the particular needs of the patient with DM. Patients with decreased pulmonary function may require special arrangements postoperatively, and severe muscle atrophy can affect the dosing of anesthetic drugs. DM can present many complications, and the anesthesiologist must take great care in the preoperative assessment of the patient.

References 


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Lesson 235: Preanesthetic Assessment Quiz

Select the single-letter response that most correctly answers the question or completes the sentence.

1. Which statement is true concerning the epidemiology of dermatomyositis (DM)?
   a. Throughout the world, incidence rates for DM in females outnumber those in males in a ratio of 2:1.
   b. The incidence of DM is about 1 per 1 thousand in North America.
   c. A seasonal onset of DM has been disproven because people are subjected to ultraviolet irradiation year-round.
   d. In general, adult DM has the highest prevalence of all the idiopathic inflammatory myopathies.

2. The patient with DM is more susceptible to all of the following except:
   a. interstitial lung disease
   b. opportunistic infections
   c. viral infections
   d. cancer

3. Which of the following statements is false regarding cutaneous lesions of DM?
   a. Patients with an acute onset of skin lesions usually deteriorate quickly, and in many, the disease is resistant to treatment.
   b. The heliotrope rash and Gottron's papules are hall mark skin lesions of DM.
   c. The presence of 2 "major" cutaneous criteria indicates a cutaneous manifestation of DM.
   d. A skin biopsy typically shows epidermal atrophy resulting from perivascular inflammation.

4. In the diagnosis of DM, the most important laboratory test is considered to be:
   a. needle electromyography
   b. measurement of serum creatine kinase levels
   c. major histocompatibility complex class I antigen expression
   d. muscle biopsy

5. Autoantibodies involved in DM include all the following except:
   a. antibodies that deposit in the dermal–epidermal junction
   b. antibodies that inhibit translational transport
   c. antibodies that inhibit protein synthesis
   d. antibodies that cause antisynthetase syndrome

6. In the treatment of DM, the physician should:
   a. pay attention to laboratory test results because serum enzyme levels closely parallel the disease state
   b. not worry about cutaneous lesions because disease weakness is of greater concern
   c. monitor muscle strength because treatment is based on observed results
   d. rely heavily on corticosteroids because patients with unresponsive disease just need a larger dose

7. Which of the following statements is false?
   a. Autoantibodies of DM attack only muscle fibers, causing necrosis and inflammation.
   b. Many factors trigger DM, such as ultraviolet irradiation, which leads to an autoimmune response.
   c. Muscle biopsy specimens of DM patients show decreased capillary density.
   d. Many symptoms of DM can mimic those of lupus erythematosus.

8. Amyopathic DM is different from DM in that:
   a. it affects distal muscle groups, whereas DM preferentially affects proximal muscle groups
   b. it does not involve autoantibodies; rather, it arises from complement deposition at the dermal–epidermal junction
   c. patients with amyopathic DM demonstrate muscle weakness stemming from an enzyme deficiency
   d. the physician may not detect muscle weakness in patients with amyopathic DM

9. The muscle biopsy specimen of a patient with DM will most likely show:
   a. perivascular atrophy, particularly in the distal extremities
   b. dilated capillaries and a subsequent increase in the ratio of capillaries to muscle fibers
   c. inflammation and fiber necrosis in the perifascicular space
   d. lymphocyte infiltrate, most commonly CD8+ cells, in the perifascicular space

10. Extramuscular manifestations that have been observed in patients with DM include:
    a. interstitial lung disease
    b. cardiomyopathy
    c. calcifications
    d. all of the above

To receive CME credit, you must complete this form, including the time attestation and evaluation, score ≥80% on the quiz, and return the form with a check for $10 made payable to MSSM-Anesthesia before November 30, 2005, to Mount Sinai School of Medicine, Department of Anesthesia, One Gustave L. Levy Place, Box 1010, New York, NY 10029. Credit will be awarded only for lessons completed with a signed time attestation that are postmarked before the expiration date.

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Your frank and considered evaluation will be helpful in improving our CME programs. Your assistance is greatly appreciated.

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   Strongly Agree Agree Disagree Strongly Disagree
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14. If you were designing the program, would you modify the structure? If so, how?
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